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THESIS
on
THE ETIOLOGY, AND THE PATHOLOGY, OF THE CHOREA OF SYDENHAM.

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By

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March 1903.



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1.HISTORY.

From time to time, in the history of Medical Science, many different forms of disease have been classed under the name of Chorea Scanti Viti. They include forms of dancing mania, various conditions allied to hysteria, and other states of muscular inco-ordination due to degenerative lesions of the nervous system.

The recognition of Chorea Minor, as an acute disease of childhood, distinct from the other forms, ^e ~~date~~ from the time of Sydenham, who first described the disease in his *Schedula Monitoria* (1686), and in his *Processus Integri* (1693), in which he gave a brief but graphic account of the disease. He recognised its affinity with the period of growth, but of its pathology he knew little or nothing. He says, "Now this affection arises from some humour falling on the nerves, and such irritation causes spasm".

Over two hundred years have elapsed since then, and we are still endeavouring to discover its cause. It is unfortunate that Sydenham adopted the name, as Paracelsus had already designated the various hysterical forms of

(History)

which he wrote, as Chorea Scanti Viti, and to this day the disease is known to the laity as St. Vitus' Dance. The modern study of the disease dates from 1810, when Bouteille wrote a noted treatise on the disease. (*Traité de la Chorée*, Paris 1810), he classified the disease and seems to have been aware of its connection with rheumatism in many instances. He was evidently aware of the difficulties attending its study, he writes, - "Tout est extraordinaire dans cette maladie; son nom est ridicule, ses symptômes, son caractère equivoque, sa cause inconnue, son traitement problematique."

In the same year Bernt of Prague also wrote a valuable monograph on the disease. Later we come to the works of Trousseau, and See, (G. Seé, *Dela Choreé Mem. De L'Acad. De Med.* 1850), and others who demonstrated its connection with rheumatism.

Roger in 1866, upheld the theory of its being rheumatism entirely. (*Archives Generales de Medeciné* 1866).

Rillet, Grisolles, Bouchut, and Steiner, especially the latter were opposed to this. Bouchut attributed it to the reaction of anaemia.

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M. Charcot, (Lecons du Mardi 1887-88-89), upheld that rheumatism was the chief cause.

Bright, in vol. 2 of the Reports on Medical Cases 1831, writes on chorea, and its connection with rheumatism, and draws attention to its association with pericarditis.

In Guy's Hospital Reports, are papers by Babbington, (Vol. 6), 1841; by Hughes in 1846; by Hughes, and Burton Brown in 1855; by Lever in 1847; by Pye-Smith in 1874; and Goodal in 1890.

M. Comby, (Prog. Med. 1888) considered chorea as, "une nevrose de croissance", having intimate relations with hysteria.

The embolic theory as to the cause of the disease was produced about the same time by Kirk^{es}, Tuckwell, and Meynert. This was supported later by Hughlings Jackson. Octavius Sturges has maintained that the disease is a purely functional one.

More recently several writers have asserted that the disease is of an infective character. Triboulet deals extensively with this subject in his, *Du Rôle Possible de L'Infection en Chorée*.

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Pianese, Berkley, and Dana, have also written on the same subject.

Of other authors who have contributed extensively to our clinical, and anatomical knowledge of the disease may be mentioned, Begbie, Ogle, Broadbent, Duckworth, Gowers, Gee, Money, S. Mackenzie, Wood, Herringham, Jacobi, Weir Mitchell, Sinkler, Lewis, and others.

2. Etiology.

AGE. It is a remarkable fact that the greatest number of cases of chorea develop at the period, when the growth of the child is the most active.

Stephen Mackenzie gives statistics of age in 439 cases, as follows;-

34% occurred between the ages of 5 & 10 years,

43% " " " " " 10 & 15 "

15% " " " " " 15 & 20 "

leaving only 8% occurring before the 5th. year and after the 20th. year. Those cases which occur after the 20th. year, are probably mixed neuroses occurring with other disturbances amounting in some cases to insanity, or dementia. In Mackenzie's cases the largest number were at the 13th. year. In the statistics of M. Germain Séé furnished from 531 cases during a period of 22 years, at L'Hôpital des Enfants Paris; we find as follows,-

28 children were less than 6 years of age,

218 " " from 6 to 10 years of age,

235 " " " 10 " 15 " " "

M. Séé remarks, "Que cette statistique ne comprend pas le

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le début réel de la maladie mais plutôt la période d'état parce qu'elle repose uniquement sur les registres de l'hôpital et que les malades ne viennent réclamer des soins que lorsque la maladie a déjà une certaine durée".

He however gives statistics of 191 cases where a careful enquiry had been made; they are as follows,-

11	were	less	than	6	years	of	age,
94	were	from	6	to	11	"	"
57	"	"	11	"	15	"	"
17	"	"	15	"	21	"	"
12	"	"	21	"	60	"	"

He adds that the maximum is reached between the ages of 6 and 11 years of age. (*Essai de Pathogénie de la Choroë de Sydenham. Duchâteau. P. 45*)

Osler out of 522 cases gives the following statistics,-

1st.	Decade	261	cases,
2nd.	"	248	"
3rd.	"	10	"
4th.	"	1	"
Above 4th.	2	"	. (<i>Osler. Practice of Med. Clinica</i>)

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West gives statistics of 1121 cases as follows,--

Up to 5 years of age 111 cases,

Between 5 and 9 years of age 510 cases,

Between 9 and 10 years of age 500 cases,

Barthez, and Sanne, give statistics of 334 cases as follows,

Between 3 & 6 years 23 cases,

" 7 & 10 " 134 "

" 11 & 14 " 169 "

" 15 & 17 " 8 " . (Buchateau. p. 45)

From these statistics it appears that chorea has two maximum periods of frequency, viz.

1st. between 7 & 9 years of age,

2nd. " 14 & 16 "

It is very interesting to note that these maximums correspond to the periods of most active growth.

The investigations on growth were published in 1881 by Dr. Hertel. He measured, and weighed 1500 school boys, and 3000 girls in schools, he gives the following statistics,--

Between 7 & 8 years, the growth was the greatest,

" 9 & 13 " the increase is less,

" 14 & 16 " the time of puberty, the child

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increases very quickly both in size and weight, and that these periods were more marked among the girls than the boys.

Merckling-Hauser, the director of the Institution for deaf mutes at Copenhagen, has also made investigations as to which period of the year the most growth takes place. He states that children grow least from the end of November, to the end of March. Much in height, but little in weight from to July and August; much in weight ~~in weight~~ from August to November.

The fact that this disease so often shows itself, just when the whole body is an extreme state of activity, owing to the increased growth, and development, is an extremely interesting etiological fact. It is at this time that the higher mental centres are beginning to be educated.

M. Comby says, "C'est en effet avec la croissance que coincide la suractivite cerebrale et medullaire c'est l'age ou les enfans prennent gout a l'etude, ou l'imagination s'eveille, avec les sentiments affectifs, ou certaines lectures font entrevoir au collegien des

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horizons inconnues, son imagination s'y plonge avec avidité et c'est le système nerveux qui paie tous les frais de cette débauche cérébrale; C'est à ce moment que les fonctions de la génération s'établissent." (France Médicale, 1888).

At this time therefore the nervous system must receive increased nutrition to enable it to carry on its increased work. If as so often happens, the child at this time suffers from some condition, which affects the proper nutrition of the body, and the nervous system in particular such as; anaemia, dyspepsia, rheumatism, etc. that very system must suffer, being in a debilitated condition it more readily, (especially if there is a predisposition) becomes affected by such diseases as chorea. We often observe children to suffer in many different ways at this period of active growth, without having any definite disease. They often grow in height out of proportion to their weight, their habits and memory become affected in various ways, often any exertion seems a trouble to them. They often complain of sleeping badly. In others it is the appetite that is affected, in some it is

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small or absent altogether; whilst in others, it becomes ravenous. All these may lead to a faulty condition of nutrition, and probably to a defective elimination of waste products. Especially ⁱⁿ the condition of fatigue, which the fast growing child so often complains of.

In all conditions of fatigue, even in health, there exists in the blood, a great quantity of waste products and toxines.

Experiments have been made on animals at rest, by injecting them with the blood of animals suffering from extreme fatigue, and have caused death, with marked toxic symptoms.

M. Bouchard in his, *Lecons sur les Auto-Intoxications*, says, "that among people who work intellectually, the urine is twice to four times more poisonous, than when taken during a period of sleep." Then we come to the conclusion that any condition which affects the nutrition of the child, especially at these periods of most active growth, may indirectly lead to a condition suitable for the development of chorea. The strain of modern education seems to be an important factor in the etiology of the

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disease, and Sturges has called particular attention to what he has designated, "school made chorea".

SEX.

Most authors agree that sex is an important etiological factor; and that the female sex is the most prone to the disease. Certainly statistics point to this conclusion. This is said by some to be due to the lack of development of the inhibitory power of motor centres, especially at the beginning of womanhood.

Statistics. Out of 554 cases at the Philadelphia Infirmary, 71 per cent, were females, and 29 per cent males. After puberty the percentage in females increases. Gowers, found 365 boys out of 1000 cases.

West, out of 1121 cases, found 347 boys, and 774 girls.

Barthez, and Sanne, in 326 cases found 81 boys, and

245 girls. Handford gives statistics of 154 cases

that had been in the General Hospital Nottingham

during a period of 14 years. The males were to the

females as 1 to 2.4 before the age of puberty; and as

1 to 4.5 after that age.

Garrod says out of 80 cases, 61 were females.

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Hughes, Steiner, Smith, Leroux, and Triboulet, are all unanimous in their statistics, that there are nearly two girls, for each boy, in cases of chorea.

Seasonal Relations.

Gerhard, Mitchell, and Lewis have made a number of observations upon the relative frequency of cases of chorea at different seasons of the year. The last named author in his paper entitled, "A Study of the Seasonal Relations of Chorea, and Rheumatism, for a Period of 15 years"; analyzes as regards the months of onset in 717 cases of chorea. His results are as follows;

"November shows the fewest attacks, viz. 24, or 3.3%.

December, 56, or 7.8%. The number of cases remains almost stationary during January, and February, then suddenly increases, reaching the highest point in March, viz. 101 cases or 14%. In April a fall occurs to 63, or 8.7%. then a rise takes place in May to 80 cases or 11.1%, then gradually falling to its lowest point in November.

Lewis' studies include a careful comparison of the

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frequency of attacks of chorea with the mean relative humidity, the mean barometer, the mean daily range thermometer, and the amount of sunshine or cloudy weather. The conclusions drawn by Lewis from his studies which extended over a number of years are as follows. -

1st. "The seasonal relationship of chorea, and rheumatism is proven".

2nd. "There is a marked resemblance in form between chorea and rheumatism tracings, and the tracing representing the total amount of sickness present in the community per month."

3rd. "This monthly variation in amount of sickness is not a cause in the fluctuation in the chorea and rheumatism tracing, but is itself probably due to the same influence."

4th. "While over study assuredly plays a most important rôle in predisposing children to chorea, the months of greatest study, and therefore, presumably of the greatest depression of bodily vigour, do not coincide with, or even precede with any regularity, the months of greatest frequency of the disease."

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5th. "It is more than probable that weather is one of the most important predisposing causes,****".

6th. "Either this apparently close relationship must be acknowledged to have an important place in the etiology of these diseases, (chorea and rheumatism) or else the resemblance must be considered to be purely accidental, which seems most unlikely from a study of the statistics shewn."

3. DISTRIBUTION OF CHOREA.

***** " *****

Station of Life.

The disease is far more common among the children of the poor, than among the children of the rich. More particularly the children of the working masses in the great centres of industry. In the report of the Collective Investigation Committee of the British Medical Association 1887^{it}, gives a percentage of 72.27% of cases as belonging to the lower classes. Improper feeding, (which is such a great cause of many diseases in this class), neglect, ill-usage, insanitary dwellings, along with the accompanying weakness, and anaemia, are frequently antecedents to the disease. We however also find it where the surroundings of the patient are all that could be desired.

Locality.

In the Report already mentioned of the British Medical Association, on the Geographical Distribution, of rickets, acute, and sub-acute rheumatism, chorea, cancer, and urinary calculus, in the British Islands, and published

(Distribution)

in the British Medical Journal, Jan. 19, 1889, in the summary of an exhaustive report on their distribution we find the following remarks on chorea.

"That Chorea, like rickets, is mainly a disease of towns and industrial regions, though by no means unknown in agricultural and rural districts; that it is fairly evenly distributed in the rural districts of the four countries; that it is rare in sea side watering places; that it is, by exception, somewhat rare in the south as well ^{as} of the west of London, and not generally speaking, common in Glasgow, even in the poorer quarters. That the distribution of chorea is further affected by that of acute and sub-acute rheumatism, its prevalence diminishing as the latter disease becomes rare". This is what one would naturally expect, the very same conditions of life as regards, hygienic surroundings, mode of life, feeding, etc. which predispose to chorea, is attributable to rheumatism as well. In fact these conditions seem to be predisposing causes to many of the diseases of childhood. Why however certain large centres of population should

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show an exemption contrary to the general rule is more difficult to explain.

Race.

It appears chiefly to attack the white races. Bogges in all his wide experience says, he never saw a case amongst negroes; although isolated cases have been described in the States of America from time to time.

4. HEREDITY. etc.

Statistics prove that predisposition is an important cause in the etiology of chorea. There is in certain cases a particular susceptibility of the nervous system, a special condition tending to the development of chorea, when it comes under the influence of an exciting cause. The part played by heredity in these nervous diseases is defined by Professor Raymond in the following words: "L'Hérédité nerveuse est l'aptitude à faire eclorre des affections nerveuses, conférée à un organisme vicié dans ses caractères anatomiques apparents ou dans les deux à la fois, par des son fonctionnement psychique ou dans les deux à la fois, par des générateurs placés dans les mêmes conditions d'hérédité ou soumis à certaines influences pouvant agir sur le système nerveux, telles que l'alcoolisme, la syphilis, la tuberculose. etc." (Hérédité Nerveuse, Bulletin médical, 3 Avril 1895).

M. Blocq, on heredity in chorea says as follows. -

"Dans la chorée de Sydenham, on trouve rarement l'hérédité similaire (père ou mère choréique, enfant choréique), et le plus souvent, il s'agit d'hérédité nerveuse dite de

(Heredity)

transformation (parents épileptiques, ataxiques, neur-
asthéniques, etc.). L'hérédité directe qui est le facteur
primordial de la chorée de Huntigdon, a même été mise en
doute par M. Raymond; il en existe actuellement cependant
des exemples relativement nombreux". (Traité de Médecine,
publié sous la direction de MM. Charcot, Bouchard, et Briss-
-aud.^{"Chorée"}) That there is a tendency to the disease in certain
families seems proved. Now and then we find that the
mother of a choreic patient has suffered from the same
disease when a child, in one instance mentioned by Osler,
both mother and grandmother had suffered from the com-
plaint. It is not however a frequent occurrence. Triboulet
only found an hereditary history four times out of 300
cases (Paris Thésis. 1893). From statistics of other
authors, it seems that about 10%, to 12% of cases can be
traced to an hereditary, or predisposing influence.
Germain Seé found 18 such histories out of 128 cases.
Herringham 12 out of 75 cases. Money, 14 out of 214 cases.
Pritchard 15 out of 125 cases.

(Heredity, etc.)

Temperament.

It is generally admitted that this is an important rôle in the etiology of chorea. Dull, heavy, phlegmatic children seldom suffer from chorea; while clever, high-strung, neurotic children seem specially prone to the disease.

Reflex Irritation.

Many of the older writers have laid great stress on the so-called reflex chorea, which was thought to be due to digestive troubles, the presence of intestinal worms, and various genital irritations. Modern writers however do not recognise any relationship between these conditions and chorea. Straton, (B.M.J. 1885), lays great stress upon the fact that erosions of the mucous membrane of the nose and throat by affording means of entrance to micro-organisms, might be the indirect cause of chorea in children. In every case that has come under my notice, caries of the teeth have been present.

Psychical Influences.

These also have always been held to play an important part in the causation of the disease. Fright, trouble, sudden grief, religious excitement, or the strain of

(Heredity) (etc.)

modern education have all been from time to time assigned as causes. In the majority of cases no very close connection between the fright, etc. is found. The disease being well known to the laity under the name of St. Vitus' Dance, and in many places even to-day, mixed up with a considerable amount of superstition; parents and friends are generally ready with such a history, and attribute the disease to such a cause. In 86 cases given by Osler in his work on Chorea, fright was said to be the exciting cause, by the relatives or friends. He however says; "In the majority of these no very close connection existed between the fright, and the onset of the chorea, as usually an interval of two or more days had elapsed". (*Chorea*, 6th ed. p. 10)

Infectious Diseases.

fever
Scarlet, may be a direct antecedent, Sturges states that a history of whooping cough prior to the attack of chorea is often met with. It is sometimes met with in the course of many of the septic diseases, such as puerperal fever, pyaemia, and multiple suppurative polyarthritis.

Cases have been reported as occurring after diphtheria,

(Heredity, etc.)

measles, scarlet fever, and typhoid fever. On the other hand, Trousseau, Radcliffe, West, Rilliez, and Barthez, all report cases where the chorea has become suspended on one of these acute diseases becoming developed. It is however interesting to note that canine chorea, often follows an acute infectious disease. Strumpell, says that chorea often develops after the infective fevers. M. Triboulet who has upheld the infective theory of chorea; out of 300 cases, says, he found 100 cases where the chorea developed during the course of one of the general febrile disorders, scarlet fever, measles, whooping cough, small pox, pneumonia, typhoid fever, intermittent fever. (placed in the order of most frequency).

Traumatic Influences.

Falls, and contusions of the head, have been mentioned where chorea has followed. Sexton mentions a case where chorea followed an operation for a severe fracture of the humerus. Sinkler (Pepper's System of Medicine), mentions a case that followed a minor surgical operation. It seems doubtful whether these recorded cases were the true chorea of Syden-

(Heredity, etc)

-ham; or due to lesions of the cortex. One case that came under my notice, the child developed choreic movements on the right side, after a fall on the head, which passed off in a few days without treatment.

Relation to Eye-Strain.

It has been claimed by Stevens, and others; that ocular defects lie at the basis of many cases of chorea.

De Schweinitz, (New York Med. Journal, 1887) made a number of investigations to test the truth of this. He found that ocular defects do not occur more frequently in choreic children than in others, although when occurring and remedied, materially affects the course of the disease.

5. CHOREA, AND PREGNANCY.

***** " *****

Chorea in pregnancy is a relative infrequent complication. The disease is of most frequent occurrence in primigravidae and usually makes its appearance in the first half of pregnancy, and is liable to recur with succeeding ones.

The facts at present known with reference to the causation of the chorea of pregnancy are briefly as follows;-

1st. There is usually a predisposition to the disease inherited or acquired.

2ndly. Inanition of the central nervous system incident to the hydraemic condition of the blood in pregnancy.

3rd. Also various potential peripheral irritants in connection with the sexual organs have been described.

Wood, in his Nervous Diseases, 1887, says, "the most rational explanation of the chorea of pregnancy is that it varies in its immediate pathology, the pregnancy simply producing a condition of the nervous system which predisposes it to be thrown into active chorea by various exciting causes".

Speigelberg, and Fehling give statistics of 137 cases with a death rate of 28%.

(Chorea, & Pregnancy)

Buist before the Edinburgh Obstetrical Society June 12th. 1895, gives statistics of 255 cases, "66 had suffered from chorea previously to the pregnancy, in 6 cases the chorea appeared at the onset of the pregnancy. As regards age the statistics shew that it becomes increasingly less common after 24 years of age. With reference to the period of pregnancy when the symptoms began, it was found that 108 cases commenced in the first three months, 70 during the second three months, 25 in the last three months. There were 45 fatal cases in 255 pregnancies where this condition existed, giving a death rate of over 17.5%. This is probably much in excess of the actual mortality, as minor cases are not reported, and some of these deaths are due to other causes, and complications, such as eclampsia, albuminuria, etc. Sixty one cases recovered before the onset of labour, 91 after the birth, and 7 became chronic. It was interesting to note that many of the cases escaped entirely in subsequent pregnancies."

Prof. Krafft Ebing, (Allg. Wien. Med. Ztg. 1897), says chorea gravidarum occurs generally in primiparae who have married young, and are of weak constitution, and occurs as a rule

(Chorea, & Pregnancy)

after the fifth month. Bamberger has shewn that there is a chorea gravidarum gravis which commences in the eight month or the ninth month, with a mortality of 30%.

I have seen two such cases commencing in the eight month, both cases were severe, and had suffered from the disease in childhood.

Pregnancy is prematurely interrupted in about one half the cases. The child may be born alive, and in some cases affected with the disease. Sinkler reports a case of congenital chorea, said to be due to fright of the mother during pregnancy. Mania, loss of memory, grave cerebral lesions, and spinal lesions, are occasionally traceable to this disease during pregnancy. Prof. Simpson, is of opinion that the high mortality in these cases, is due to the fact that the induction of labour is often too long delayed.

6. CHOREA, and its CONNECTION with

other NEUROSES.

***** " *****

Some authors have described a connection between chorea and other neuroses, and that the symptoms of these latter may accompany, or complicate the former. The diseases especially mentioned are hysteria, epilepsy, ^aparalysis agitans, and paralysis. It is undoubted that patients suffering from chorea often exhibit frequent emotional disturbances of the mind which may be regarded as hysteroidal.

Whether these are the results of the tormenting nature of the disease they are suffering from, or of a true hysteria, or not, it is very difficult to say. Charcot is an upholder of the affinity that exists between chorea, and hysteria.

He says;—"Certes il existe entre la chorée et l'hystérie une grande parenté de même qu'entre la chorée et le rhumatisme,

de même encore qu'entre le rhumatisme et l'hystérie". *(Duchâteau
Paris, 1893.
part. 16)*

Certainly it is a fact worth recording that one often finds

a family history with some trace of hysteria in that of chorea patients, just as one often finds a rheumatic family history amongst hysterical patients. Emotion which plays such an important rôle in hysteria, is often the only cause to

(Chorea, & other Neuroses)

which we can attribute an attack of chorea. The French school, and especially Charcot have recognised the fact that the condition known as hysteria may affect even quite young children. When it occurs in infants the cause is, according to the English school, due to hereditary neurosis. Hysterical affections are often found at an early age in children of a neurotic disposition. In hysteria, in children, Clouston says,—"There is no doubt a pathologically premature development of the emotional brain centres, due to a bad heredity, and this is sometimes accompanied by a premature sexual development." (*Neurosis of Development, Clouston.*)

In connection with chorea, clinical facts seem to indicate that a certain proportion of cases are a kind of pseudo-chorea, hysterical in origin. In connection with this subject, an interesting case came under my notice some years ago. A child age 4, was occupying the same room as her sister age 7, who was suffering from chorea. She also developed marked symptoms of chorea. On being isolated the symptoms disappeared in a day or two. This child suffered from an ^{on-}unrollable temper, and had had acute rheumatism.

(Chorea, & other Neuroses)

More serious psychoses are occasionally observed, although they appear to be rare. Active Mania has been seen in grave choreic cases, as well as states with hallucinations.

Evan Powell gave an account before the Nottingham Med. Chir. Society Dec 7. 1888, of two cases complicated with insanity as a result of the choreic affection. The first was that of a young man, who had suffered from rheumatism, and great mental anxiety, this was succeeded by chorea, followed by insanity. Death resulted from exhaustion.

The second was that of a young woman, age 20. She had a good family history, no taint of rheumatism, heart disease, or nervous affection. She had never suffered from any illness until six weeks before her admission to the Nottingham Asylum, when the catamenia did not appear, at the proper time, and she became much distressed; she placed herself under medical treatment, and in three weeks had, what was described as, "an hysterical fit", four days after choreic movements began in her limbs and face, which gradually became more violent, and two weeks after symptoms of insanity commenced. She became maniacal, and had hallucinations of sight, and hearing. The spasmodic movements

(Chorea, & other Neuroses.)

were general. There was no abnormal cardiac sign, except that the beat was rapid 140 per minute. The choreoid movements gradually left her limb and trunk muscles, but continued in those of the tongue and pharynx, so much so that swallowing and speech were rendered very difficult; her mind also became clear, and calm. She gradually sank from exhaustion five days after admission. On post mortem the brain was found intensely hyperaemic, but no other disease was made out. These cases show how important a part a mental anxiety may play in the causation of chorea. Paralysis Agitans does not appear to play an important part as a result or a complication of chorea. Paralysis, in varying degrees, is an undoubted accompaniment or sequela of chorea; whether directly caused by the same lesion of the central nervous system, or simply by the over-action of the convulsed muscles is an open question. That mental and emotional impressions are often exciting causes of the disease we are bound to admit. Like hysteria the disease is mostly confined to the emotionally impressionable sex. It is a disease like hysteria of towns and

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of town life, and of civilised races. It is often found in phthisical families, but still oftener in families with a history of epilepsy or insanity. Neurotic heredity certainly plays a great part as a predisposing cause of the disease; a combined neurotic and rheumatic heredity being its very strongest predisposing cause. Even in children it is usually accompanied with mental disturbances of a mild kind, such as inco-ordinated mental action, hallucinations of hearing, in some cases severe and forming a distinct form of insanity. In others we see disturbances of feeling so common in hysteria.

The relation of chorea to epilepsy is denied by some authors, while others uphold the relationship. The weight of opinion seems to be in favour of it. The acute form of the disease gives support to this view. Prof.

Charcot in his description of acute chorea, draws an analogy to the form of epilepsy known as the status epilepticus, a view which has much to be said in its favour, for the conditions are very similar, both in the violence of their movements and in their fatal terminations. That mental conditions in children play

(Chorea, & other Neuroses)

this part is explained by Dr. Sturges in his, "Chorea and other allied Movement Disorders of Early Life", he says, - "The muscles concerned having been educated more or less perfectly in certain kinds of movement, are suddenly thrown back in their education by some nervous shock which renders them restless and unruly, while the operation of the will, no longer sufficing to regulate the limbs, tends rather to disfigure the movements it seeks to arrest. Intelligent movement is for the while embarrassed, disfigured and overborne by emotion movement."

At the same time the various physical causes which are undoubtedly contributory must not be forgotten at this period of life, and not infrequent association of impaired health, from many of the diseases incidental to this time of life, either antecedent or co-incident, may readily be supposed to influence the nutrition of the nervous system, rendering it more irritable and liable to various nervous affections.

The study of the pathology of these neuroses has, as yet, taught us very little as to their exact nature, the

(Chorea & other Neuroses)

the clinical history has established certain facts. The nervous system of all others seems to be affected by what we term heredity or predisposition. Children notoriously resemble their parents, not only in their general configuration of body, features, etc. but also in their mental attributes, trivial peculiarities of speech, manner, laugh, and gesture; and are often handed down from one generation to another. It is therefore not surprising that morbid conditions of the nervous system should likewise be transmitted. It is important to remember that these inherited predispositions often miss a whole generation only to recur often with renewed vigour in a subsequent one. Often in cases of chorea, epilepsy, hysteria, and other nervous affections, the immediate family history shows no taint, but on enquiry, we find that members of the preceding generation suffered from the same disease or some kindred nervous affection.

Certain it is that these neuroses seem to run in families and are apt in some degree to alternate, so that we see a parent who suffered from hysteria, begetting children who

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suffer from chorea, epilepsy, and other diseases peculiar to the nervous system. What this affinity, predisposition, or hereditary tendency is we are unable to say; that it exists not only in the human being, but also in animals is certain. We therefore call it the Neurotic Diathesis, and we have already remarked that when this is combined with the so called rheumatic diathesis, we have then a most suitable soil for the development of chorea.

Whether we shall ultimately find that there is a common cause for these neuroses, and that there is simply a different portion of the nervous system affected in these different diseases we are at present unable to say. Dr. Radcliffe says—"How far the occurrence of chorea implies a tendency to other disorders of the nervous system, especially to epilepsy, is a question which has not hitherto been fully entertained, and I cannot supply an answer from actual statistics. But this I may say—that I have frequently met with epileptic patients who were choreic at one period of their life, and that the impression left on my mind from what I have seen is, that the chances of chorea being followed sooner or later by

(Chorea, & other Neuroses)

some other disease of the nervous system are too much made light of." (Reynolds's Syst. of Med. vol. 2 p. 125 1st. Ed. Art. "Chorea" by C. Radcliffe.)

The Exciting Causes of Neurosal Seizures.

Another similarity in the history of the various neuroses is the striking affinity one with another in the variety and similarity of the influences which operate as exciting or accessory causes of the seizures. It constantly happens, so it seems, that a malady of this class is, as it were, latent, until some disturbing influence supervenes, which ^{is} a sufficient cause to upset the unstable equilibrium of the nervous system, and bring about the actual disease which has been silently threat^{en}ing. This is well seen in Megrin, Epilepsy, Asthma, Insanity and many other similar diseases. Fright which as we have already remarked is the most commonly assigned cause of chorea, and although its influence is undoubtedly greatly exaggerated, yet Dr. Hughes saw no reason to doubt its influence in a great many cases, (Cases of Chorea, by H. Hughes Guy's Hospital Reps. 2nd. ser. V. 4. p. 374;)

Emotion certainly is important in the disease when developed, and will serve to aggravate the condition, and bring on a severe paroxysm of convulsive movements. The influence of fear, as an exciting cause in children need not surprise us, as it is one of the most frequent and serious emotions to which children are subject to. Again the neurotic disposition tends to greatly exaggerate the tendency to particular emotions. A child, for instance, with a predisposition to chorea, or epilepsy, may be more easily moved and experience a much more violent emotion from a given cause of fright than another. That heredity ^{as} transmission is of frequent occurrence in these neuroses is certain, but in addition a similar transformation is occasionally met with in the same patient, one form of seizure being temporarily or permanently ^{replaced} by another. This is, no doubt, exceptional, but occurs often enough to exclude the notion of a mere accidental succession. Prichard recognised this fact at the beginning of the last century, he writes, -

"It would appear that many of the disorders of the nervous system are more nearly connected with respect to

(Chorea & other Neuroses.)

their causes and morbid conditions in which they consist, than most of the disorders that are classed together in other departments of nosology. This remark will be illustrated by observing the mutual relations of some of these disorders, their successions, and their conversions into each other." (Diseases of the Nervous System, Pt. 1. pp. 58 & 66. Prichard, 1822).

The two cases mentioned above where the choreic condition was replaced by insanity, illustrate this statement.

Also the case of the child who developed the disease through being in constant company with her sister who was suffering from chorea, was certainly hysteroidal in origin. What the condition is that binds these various neuroses in such close relationship it is impossible to define, and therefore they are classed as of the diseases known as "The Neuroses."

7. CHOREA, AND RHEUMATISM.

***** " *****

Most authors have recognised a relationship between chorea, and rheumatism. The English and French schools maintain the connection of the two diseases.

Bouteille, in the 1st century, (Opus Cit. p. 297), draws attention to it. Bright, in his Syllabus, or Outlines of Lectures on the Practice of Medicine, published at Guy's Hospital in 1802, says that rheumatism was a recognised cause of chorea; and again in 1820 he says that, "chorea sometimes alternates with rheumatism".

Copeland also pointed out the connection with rheumatism. He writes, (London Med. Rep. Vol. 15), "The association of this disease with rheumatism has been observed by me on several occasions, and in nearly all there has been a marked disposition of the rheumatic affection to recede from the joints or extremities, and attack the internal fibro-serous membranes, as those of the cerebro-spinal axis, and the pericardium".

Pritchard, (London Med. Rep. 1824), speaks of the connection of the disease with rheumatism.

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Bright, (Med. & Chir. Soc. Transactions Vol. 22, 1839), also speaks of the connection.

G. Seé, (Mem. del' Acad. de Méd. T. 15, 1850), says, "of two rheumatic infants at least one will be choreic, and of five choreic children there are two rheumatic". He states that out of 128 cases, 61 were complicated with rheumatism i.e. 19%.

M. Roger, (Arch. Gen. Méd. 1866, & 1867), looks upon the disease as a modified rheumatism, and regards the condition as a manifestation of rheumatism. He says, "Une enfant choréique est toujours menace de rhumatisme de même qu'un enfant rhumatisant est toujours sous le coup d'une chorée."

He considers that articular rheumatism, chorea, and heart disease, were three terms of the same condition, "la chorée rheumato-cardiaque."

American and German authors have not looked upon it in the same light, although admitting a connection.

Steiner of Prague, (Dict. Dechambre, Raymond-), on the other hand says that out of 252 cases, rheumatism only complicated four cases.

Hughes, and Burton Brown, (Guys' Hospital Reports, 1856),

(Chorea, and Rheumatism.)

give 104 cases of chorea with 89 complicated with rheumatism, i.e. 85%.

Kirk⁸⁵, (Med. Times and Gaz. 1863), out of 36 cases of chorea 33 were complicated with rheumatism, and cardiac affections i.e. 92%.

Roger, (Arch. Gen. Méd. 1867 & '68), out of 71 cases 100% suffered from rheumatic affections of the heart.

Haven, (Boston Med. & Surgical Journal 1881), out of 200 cases of chorea 42 were complicated with rheumatism, i.e. 21%.

C.W. Townsend before the American Pediatric Society at Boston, (Trans. of the Amer. Ped. Soc. Vol. 4 1892), reported 148 cases with a history of rheumatism in some form either before or after the chorea, in 54% of cases.

M. Allen Star, Gives statistics of 385 cases, with a history of rheumatism in 18%.

Garrod, (Royal Med. & Chir. Soc. London 1889), out of 80 cases 40% had a rheumatic family history. In 15 there was a personal history of rheumatism with fever, in 9 of rheumatism with swollen joints, and in 9 of joint pains only.

(Chorea, & Rheumatism.)

Herringham, (Royal Med. & Chir. Soc. 1889), out of statistics of 80 cases of chorea, says acute rheumatism preceded the attack in 19 cases, immediately in 4, at some interval in 15. It accompanied the chorea in 2 cases. Pains in the joints preceded the attack in 15 cases, accompanied it in one case. The total number in which rheumatism could be traced was 37. Family History, (calculated from parents, brothers, and sisters only), rheumatism had occurred in 25 out of 75 families.

Osler, out of 554 cases, says in 15.5 there was rheumatism in the family history, in 15.8 there was a history of articular swelling, and in 33 cases there was a history of pains described as rheumatic in various parts of the body. If these are regarded as rheumatism we have a percentage of nearly 21%.

In the Report of the Collective Investigation Committee of the British Med. Assoc., out of 439 cases investigated 22% had had rheumatism with distinct joint affection.

Meyer, (Polyclinique de la Charite, Berlin), out of

(Chorea, and Rheumatism.)

121 cases says 20% were complicated with a rheumatic history.

Goodall, (Ibid 1890), out of 262 cases 32% were associated with rheumatism.

Sturges, (Lancet, 1883), gives 202 cases of chorea, 38 of which had a history of rheumatism.

Prior, (Berlin Klin. Woch. 1886), gives 92 cases with a history of rheumatism in 5 only.

Comby, (Tr. Méd. 1888), gives an account of 16 cases none of which had any history of rheumatism.

Leroux, (Rev. Mal. Enf. 1890), gives 80 cases with a history of rheumatism in 5 cases.

In considering these statistics we must always bear in mind that for the most part they are taken from hospital reports, where naturally we have the worst type of cases to deal with.

Sturges says, (Lancet 1883), that among adults in hospitals the proportion of cases with rheumatic histories, varies from 12% to 20%. Among children the percentage is 15%.

The fact that many cases have no rheumatic history, personal, or family, and that only a certain percentage

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of cases show it,makes it more than ever questionable whether these so called rheumatic complications of chorea are true rheumatism,or similar to those we see in gonorrhoea,and other septic conditions,which are not regarded now as true rheumatism. The rheumatic pains that we get in gonorrhoea,are little affected by the specifics used in acute rheumatism,and seem to be due to an infective process due to the presence of the gonococci or their toxins in the blood.It is not unreasonable to suppose that these joint pains of chorea may be due to a similar cause.That the rheumatic diathesis is a favourable one for the development of the disease we are bound to admit,but that does not prove the disease to be rheumatic in origin.

Another slight argument in favour of these pains not being true rheumatism,is shewn in the statistics of acute rheumatism.Here we find that adults between the ages of 16 & 25,and the male sex are most prone to the disease.Now we have shewn by the above statistics that chorea has a special affinity for children,and the female sex.

In hysteria,epilepsy,and neurasthenia,we often find

(Chorea, & Rheumatism.)

them complicated with rheumatic affections of various descriptions, yet they are always looked upon as distinctly nervous diseases. M. Rilliet says that in Geneva rheumatism is very common, and that chorea is very rare. In the statistics given we find that 795 cases suffered from various rheumatic affections out of a total of 3102 cases, or 25.3%. In many of the statistics it was admitted that a considerable number of the cases only suffered from vague pains in the joints. Even admitting these to be rheumatic in origin, the percentage is not large enough to attribute rheumatism as an exclusive cause of chorea. All that we can admit is that the condition known as the rheumatic diathesis is one favourable to the development of chorea. Again the heart complications have in some statistics been all classed as rheumatic in origin, which leads to another fallacy in coming to a correct conclusion as to the true rheumatic percentage.

8. ENDOCARDITIS, AND PERICARDITIS,

IN CHOREA.

***** " *****

Acute endocarditis, commonly of the mitral valve, occurs with great frequency in chorea and it seems justifiable to say that there is no disease, not even acute rheumatism which is so frequently accompanied with endocarditis. Osler, says, "The extraordinary frequency with which mitral valvulitis is met with in fatal cases of chorea is remarkable. There is no known disease in which endocarditis is so constantly found post mortem as chorea; it is exceptional to find the heart healthy. Statistics of fatal cases of acute rheumatism do not show so large a percentage of endocarditis. The lesion is identical microscopically as well as macroscopically, with simple or warty endocarditis as we see it in other diseases." (*Chorea* Osler p. 50)

As we have already remarked in speaking of rheumatism, the heart complications of chorea, viz. endocarditis, and pericarditis, have been classed by some authors, as evidence of rheumatism; now we know that in the ulcerative form of the disease, micrococci of various forms, chiefly

(Endocarditis, & Chorea.)

Staphylococcus pyogenes aureus, and *Staphylococcus pyogenes*, have been found.

Orth, and Wyssokowitz, (Central B.F.D. Med., Wis. 1885. No. 33.), experimentally produced ulcerative endocarditis by injuring the aortic valves and then injecting staphylococci into the blood.

Ribbert, (Fortschr. d. Méd. 1886) (No. 1), produced endocarditis without injuring the valves.

In fatal cases of gonorrhoeal rheumatism with endocarditis, the gonococci have been found on the vegetations on the valves. Pneumonococci have also been found. In fact endocarditis, and pericarditis, are common complications of all the infective diseases. It is therefore certain that this condition cannot be looked upon as rheumatic in origin entirely, and therefore the statistics, (as we have already remarked) of observers who have classed all cases with heart complications of endocarditis are extremely misleading in coming to a definite conclusion as to the relative frequency and relation of chorea and rheumatism.

(The consideration of the infective nature of these diseases in connection with chorea, will be discussed

(Endocarditis & Chorea.)

later.) Again there are cases of chorea that have exhibited heart murmurs that have been attributed to endocarditis during life, and yet after death the valves have been found perfectly healthy.

In its sequel the cardiac affection of chorea has been supposed by some authors to differ from that of other diseases. Sturges, writes, "none of the injurious after consequences which attend endocarditis in its other relations are found to ensue here". This has been proved not to be the case. Osler, writes on the above quotation of Sturges, as follows,—"The examination of a large number of choreics some years subsequent to the attacks tells a sad tale to the contrary, and proves that the primary heart troubles are, in a majority of cases at least an endocarditis with its subsequent complications."

Out of 80 fatal cases collected from the various hospitals in London, only 5 were the valves, and pericardium reported as healthy.

Osler gives statistics of 72 fatal cases, of which 61 presented endocarditis. The mitral valve seemed most frequently

(Endocarditis, & Chorea.)

affected. In 40 cases they were alone the seat of the endocarditis; in 13 cases the aortic valves were also affected; in some cases the tricuspid was involved along with the mitral. Acute or sub-acute rheumatism had occurred in 31 of the cases; and in 37 it was distinctly stated not to be present. In its constant association with endocarditis chorea stands unique. The endocarditis is usually of the simple variety; post mortem the valves present the usual characteristics met with in rheumatism, and the secondary infections in febrile disorders.

Sanson, states that there are two forms of endocarditis in chorea; the ordinary rheumatic valve thickening, and the beading of the cusps with papillary elevations. S. Mackenzie, has studied the subsequent condition of the heart in choreic patients, and in an examination of 33 cases at periods of from 1 to 5 years subsequent to the attack, says, "that indisputable heart disease persisted in 60.6% of the series of chorea patients examined." Of 44 children examined by Donkin at periods varying from 2 to 12 years after the chorea, 18 had signs of

(Endocarditis, & Chorea.)

heart disease. Osler, Burr, and Sharples, have made an investigation of 140 cases that had been in attendance at the Philadelphia Infirmary for Diseases of the Nervous System since 1876; 98 were males, and 42 females the length of time that had elapsed since the attack of chorea varied from 2 to 16 years. The result of the examination was as follows;—

In 51 cases the heart was normal.

In 17 cases there was disturbance, which was functional.

In 72 cases there were signs of organic heart lesion.

This gives a percentage of a little over 51% of cases in which, following an attack of chorea, there was definite damage apparent in the heart. There was a history of rheumatism in only 9 cases, of the 51 where the heart was normal. Of these 51, 15 had had three or more attacks, 8 had had two attacks, and 27 a single attack. Of the cases presenting abnormal signs 17 were examples of functional disturbance. Of the 72 cases with signs of organic disease 30 had had three or more attacks.

In 34% of the cases there was a history of acute arthritis

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which in 7 of the cases had followed the chorea in from one to five years. In the total number of cases the percentage connected with rheumatism was 24%, or with the group of 51 normal cases, it was a little over 17%. Thus we see the greater liability to serious heart mischief in the cases with joint complications. However we have the much larger proportion of cases viz. 66% with positive organic disease examined at a date of two or more years subsequent to the attack of chorea, in which there was no history of rheumatism in any form. The commonest lesion found was that of mitral insufficiency. There were only four instances of combined aortic, and mitral disease. With regard to endocarditis in chorea we come to the following conclusions.:-

1st. That endocarditis is a very common complication.

2nd. That in the majority of cases it is independent

of acute rheumatism.

3rd. That in a large proportion of cases, it lays the foundation of organic heart disease.

Bouillard holds that the valvular lesion is of itself a manifestation of rheumatism, he says, "chez les jeunes sujets

(Endocarditis, & Pericarditis, in Chorea)

le coeur se comporte comme une articulation."

Endocarditis is certainly the most serious complication of chorea. Chorea is rarely a fatal disease, and does not often leave permanent damage to the nervous system, so far as intelligence and motor power are concerned; but the endocarditis which one so often gets as a complication lays the foundation of permanent organic disease of the heart. The fact that endocarditis is so often found in cases of chorea is a very strong argument in favour of those who hold to the infective theory of the causation of the disease, owing to the frequency with which the same condition is seen in other diseases of an infective nature.

(Pericarditis in Chorea)

Inflammation of the Pericardium in connection with chorea was first described by Bright, who states that this had long been observed by the Guy's Hospital physicians. His statement on the subject is worth quoting; "With regard to the connection between inflammation of the pericardium and chorea, when called upon last year to deliver the Lumleian Lectures at the College of Physicians, I took occasion to state that for some years I had been persuaded of the existence of such a combination, and little attention has hitherto, as far as I know, been paid to the subject, although the combination of this spasmodic disease with rheumatism has long been recognised." (*Med. Chir. Soc. Trans. Vol. XXII. p. 10.*)

Bright thought that the connection of the two was through the phrenic nerve, which communicated the irritation from the inflamed pericardium to the spinal cord. Sibson, (*Reynolds' System of Medicine.*), gives an account of pericarditis in chorea. He states that 21 out of 180 cases of acute rheumatism with affections of the nervous system had chorea. While 15 out of the 21 with chorea had pericarditis; 14 of them had endocarditis.

(Pericarditis in Chorea.)

Osler in his work, "On Chorea", says, "In 19 of the 73 autopsies in chorea which I have recently collected, pericarditis occurred as a complication, and in 17 it was associated with endocarditis. In eight of the cases there was a history of acute rheumatism. One case had sub-acute rheumatism, one rheumatic pains, while nine had not had acute arthritis. Of the nineteen cases eight were under ten years of age, eleven were in the second decade."

9. PATHOLOGY.

***** " *****

The pathology of chorea, like that of many other neuroses has derived little elucidation from the study of the morbid anatomy of the disease except of a negative character.

In many cases of fatal chorea nothing abnormal has been found post mortem.

Many of the conditions described as having been found post mortem, are probably the result of the disease, and not the cause of it.

Dr. Dickinson who made an investigation of the pathology of the disease in a large number of cases, gave the results of his work before the Royal Medical, and Chirurgical Society, in the session of 1875, & 1876.

He described dilations of the arterioles throughout the brain and spinal cord, particularly in the optic thalamus, and the corpus striatum attended with small haemorrhages. He considered the disease as due to a widely spread hyperaemia of the nervous centres. He did not describe any embolisms.

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Tuckwell has described capillary embolisms in the central ganglia, in the central convolutions, and spinal cord, along with patches of softening and minute haemorrhages.

Most observers have described the presence of endocarditis with deposits of beads of lymph on the mitral or aortic valves, or both.

Dr. Dickinson says the changes described by him were most marked in the neighbourhood of the trunks of the middle cerebral arteries, and in the posterior and lateral portions of the grey matter of the cord mainly at the upper part; and further that in all these regions the morbid conditions tend to be symmetrically arranged. He did not consider that this wide spread hyperaemia was due to any mechanical cause, as some authors have; but due to causes of two kinds. One being the rheumatic condition so often found; the other comprising various forms of irritation, mental, and reflex belonging especially to the nervous system.

In considering these changes described we must remember that it is only the severe cases that prove fatal; where there has been extreme violence in the choreic movements

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along with in many cases, endocarditis, delirium, and often other complications.

We have in fact multiple lesions just as we have multiple symptoms, and from our knowledge of the physiology of the nerve centres and from their comparison with the pathology of other diseases of the nervous system, we have to endeavour to trace which are the result of the disease, and which can be assigned as a probable cause of the disease. The point to demonstrate is that the lesion causing the chorea, is in the brain, and not the cord.

The arguments against it being in the cord have been given by Dr. Russell Reynolds, and are as follows. -

"1st. That tonic and not clonic spasms are characteristic of spinal irritation.

2nd. The degree of control over the movements retained by the will.

3rd. Their increase under emotion, and cessation during sleep.

4th. The diminished reflex action on tickling.

5th. The phenomena of hemichorea and its relations with

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hemiplegia.

With regard to this phenomena Broadbent in his article on Chorea in Quains' Dict. of Medicine, says as follows.-

"It cannot be supposed, for example, that one lateral half of the entire length of the pons, medulla, and cord can be affected without implication of the other half, which would be in hemichorea of spinal origin. Still more conclusive is the fact that when in hemichorea there is impairment of sensation it is on the same side as the movements, and not, as in hemiparaplegia (due to the division of one half of the cord) on the opposite side to the motor paralysis. The parallelism between hemichorea and hemiplegia is so perfect as to suggest at once that the two affections represent different conditions of the same nerve centres."

Flechsigs has found both internal segments of the lenticular nucleus diseased in several cases of severe general chorea with delirium.

Elischer in 1874, (Virchow's Archiv. Bd. 63), Jakovenko, and more recently Wollenberg have pointed out the existence

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of special elements which they have described as having found in the course of the capillaries of the internal segment of the lenticular nucleus, and have designated them as "les corpuscles choreiques". (Traité de méd. p. 1220 Charcot).

Turner, (Lancet, 1890) has found in the neighbourhood of the fissure of Rolando a destruction of the nerve cells, which were swollen and had lost their shape.

The well known embolic theory of Kirkes, and afterwards improved on by Hughlings Jackson is of all others the condition which might be expected to induce the instability without the abolition of function which exists in chorea. While giving the theory a prominent place, it cannot be considered the only cause.

In many cases no appreciable lesion of the nerve centres has been found.

Clouston in his "Neuroses of Development" says as follows "The most reasonable hypothesis as to the seat of the disease is, in my opinion, that the basal ganglia are at

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fault, and this occurs when the co-ordination of their motor functions with the mind are being perfected, and when their full use as motor servants and ministers, to the rapidly developing higher mental centres in the cortex, is being completed. It has always I think a neurotic heredity, even in the rheumatic cases, a combined neurotic and rheumatic heredity being its very strongest predisposing cause."

Delafond opened a great number of animals suffering from canine chorea, at different periods of the disease. He found no appreciable changes in the nervous systems, or in any of the internal organs. He found absolutely nothing conclusive; yet conjointly the symptoms of chorea are very characteristic, and always identify themselves with the nervous system. Paresis of different degrees with psychical phenomena, and the production of inco-ordinate movements, are not attributable except to the nervous system. These considerations have led Strumpell to say, "Force nous est de considérer la chorée une

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névrose, c'est-à-dire comme une maladie dont les désordres fonctionnels n'ont encore aucun substratum organique connu. La symptomatologie de cette affection dénote à l'évidence qu'elle consiste principalement en une altération d'un département moteur du système nerveux. Mais quant à savoir quel est ce département moteur spécial, on ne peut faire à ce sujet que des conjectures. Il paraît pourtant très vraisemblable que le siège propre de la chorée doit se trouver le cerveau".

(From, "Essai de Pathogénie de la Chorée de Sydenham," Paris, 1893, Duchateau) (1894, 10)

Bogges, (Pediatrics, Sep. 1898.) says,—"In treating chorea as a functional disturbance of the nervous system, we encounter the same difficulties as we do in epilepsy; yet we often find in both diseases evidences of actual cerebral disease. In post mortems upon epileptics we often fail to find gross cerebral disease when we thought it was present, and vice versa".

G. See collected 84 cases upon which post mortems had been

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made; in 16 no changes were found; in 32 there were lesions of the brain and nerve centres; in the remainder there was congestion of serous membranes.

M. Laurent, (*Traité Pratique De Médecine Clinique*, Paris 1897 p. 113), says,—"Charcot place le siège de la lésion dans la partie postérieure de la capsule interne, dans la région lenticulo-optique. Seulement la lésion porterait plus spécialement sur les faisceaux situés en avant et en dehors de ceux de l'hémianesthésie, et recouvrant l'extrémité postérieure de la couche optique." Dana, (*Brain* Vol. 13, 1890), analyzed the recorded autopsies up to that time, in 39 only were the conditions of the nervous system satisfactorily described. Out of 19 cases where minute microscopical examination had been made, in 16 there was intense cerebral hyperaemia, peri-arterial exudations, erosions, softened spots, minute haemorrhages, and sometimes emboli. Thus the vascular changes described appear to be the same as those described by Dickinson, and already referred to. No characteristic changes were met with in the medulla, or spinal cord.

Berkley has found foci of minute haemorrhages in the

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pons, especially among the fibres of the pyramidal tract. He examined the nuclei on the floor of the ventricle and more anteriorly the olivary bodies, the internal fibres of the vagus and hypoglossus, as well as the transverse fibres, with great care, but without finding the slightest pathological change in them. Likewise in the spinal cord the multipolar cells were unchanged.

In Mr. Powells' two fatal cases mentioned in the chapter on Neuroses, the microscopical examination, showed small haemorrhages chiefly in the cord and pons; but also in the motor cortex, thromboses of vessels with haemorrhage into the lymph sheath, rupture of vessels in the commissure of the cord, and one vessel in which there was, what appeared to be an incomplete embolus.

Gowers, Bristowe, Sturges, Raymond, Frerichs, Ogle, and many other authors have from time to time described similar changes, too numerous to mention here.

Osler, ("Chorea & Choreiform Affections," p. 68), in writing on the site of the lesion, says,—"That the condition found in fatal cases gives no satisfactory information on this point is evident, since, basing their arguments on these

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findings, authors have claimed in turn the cortex, the basal ganglia, and the cord as the seat of the disease. As in the consideration of the nature of the disease, the aggravated cases give us important information, so with respect to the primary change, the intense psychological phenomena indicate clearly a cortical lesion. The alterations in character, and the mental features of the malady in less severe forms, also suggest involvement of the higher centres. The cessation of the movements during sleep, and the frequent occurrence of hemi-chorea, are usually urged as favouring this view. Of course irregular movements of muscles may be produced by irritation in any part of the motor path, but "the motor impulses that excite the muscles pass to the spinal cord from the motor region of the cortex. It is here that movements are arranged, and if they are disarranged and the disorder proceeds from the brain, we naturally refer it to a disordered action of the cells of the cortex" (Gowers)."

The only just inference from this (the various theories) is, that the accurate pathology and morbid anatomy of chorea are not yet thoroughly understood. We know

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that in the majority of cases there is a considerable change in the grey-matter of the central nervous system, in the motor centres, and in the cortex.

The words of Rilliet and Barthez, written in 1867, have still to be disproved, "On peut conclure des différentes lésions rencontrées parfois dans la chorée, qu'il existe deux espèces de chorée. En un mot, il en est de la chorée comme des convulsions qui sont tantôt idiopathiques, tantôt symptomatiques." (Dict. de Méd. et de Chir. Paris, 1867). (*Chorée*)

As I have already remarked the results found in fatal cases are such that we may very well ask, are they the cause, or the result of the disease! The condition is in fact similar to an infective inflammatory process, or to one caused by their toxic products, similar to those which cause the rheumatic symptoms, and the cardiac lesions." (Dana, Text Book of Nervous Diseases, New York, 1892).

The researches of P. Manasse, (Ueber Hyaline Ballen und Thromben, In Den Gehirn Gefässen Bei Acuten Infectionen Krankheiten. Virchow's Archive, Band 130 Heft 2, p. 127,

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1890), have to a great extent proved that this is the case. He examined the brains of 39 people of which 20 were from subjects who had succumbed from the results of infectious diseases, and 19 of individuals dying from diverse non-infectious diseases. In the first series he found hyaline thrombosis in the blood vessels of the brain, which in some cases they entirely obliterated, in others occupying the centres of the vessels. He has seen in the same places almost always groups of cells, especially around these vessels. The fact that these hyaline formations did not exist in the brains of the subjects dying from non-infectious diseases, he concluded that they are produced during life, and that they are not produced by a cadaveric alteration. He experimented on dogs with intra-venous injections of septic products during life. Post Mortem examination of the brains showed the same lesions, vascular, and perivascular. He therefore came to the conclusion, that these formations and conditions are not peculiar to chorea, but belong to the infectious maladies, and are to be met with in the antecedents of these diseases.

10;THE INFECTIVE,AND AUTO-INTOXICATION

THEORIES.

The Infective Theory.

For many years past, some authors, more particularly on the continent, have tried to prove that the disease is due to a specific microbe. Owing to the fact that both the severer and the milder forms have some, and others, all the features of an infectious malady. While on the other hand many authors have denied it altogether, arguing that the psychical element in the disease is against such a theory, others pointing out the hereditary nature of the chorea of Huntington, and looking upon the chorea of Sydenham in the same light. While yet another school have adhered to the rheumatic nature of the disease, as its only cause. While the theory of infection has received much attention, and many observers according to their reports have established the infective nature of the disease, which would account for the symptoms in many cases,

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there are others where it does not.

Ossasionally one meets with a case of chorea without fever, without endocarditis, without rheumatism; here it is difficult to attribute the disease to an infection. Although on the other hand, one has not to forget that in scarlet fever, which like chorea is chiefly a disease of children and adults, the fever is often slight and un-noticed, the constitutional disturbance is often trivial, so that the disease is overlooked until desquamation appears. Such cases are of every day occurrence, yet no one would suggest that this complaint was not of infective origin.

Again certain diseases of the nervous system which were not formerly attributed to an infective origin have in recent years been proved to be so without a doubt. Take for example, Tubercular Meningitis, Cerebro-Spinal Meningitis, where the infective organisms are present in the lesions. Or Tetanus, and Diphtheric Paralysis, where the condition is due to the toxins produced by the specific microbe of the disease.

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The arguments in favour of the disease being of an infective origin, in addition to the Bacteriological researches that have been made, are as follows. -

1st. The influence of age and seasons which are common to most infectious diseases.

2nd. The clinical course of the severer forms, in which all the symptoms of a typical infective process are present.

The disease appears as it does in scarlet fever, diphtheria, and other infectious diseases in all degrees of severity, from the very slightest, to the most severe cases.

3rd. The post mortem changes such as, the endocarditis, a most distinct lesion of an infectious disease and found to almost universally contain organisms. The pericarditis, the parotitis, etc. all tend to strengthen the view of an infective process.

Even its connection with acute rheumatism in a certain percentage of cases seems in the light of present discoveries to also strengthen the infective theory

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of the disease;whether we look upon the joint affections as a distinct disease similar to, and identical with acute rheumatism, or whether we look upon them as similar to the joint affections of gonorrhoea, of scarlet fever, of septicaemia, which no one now regards as acute rheumatism.

In considering the disease as due to a distinct infection, we have to decide the two following questions, 1st. Is the disease due to a distinct specific organism. 2nd. Or is it due to the toxins produced by a specific organism, as we see in tetanus, and diphtheric paralysis. M. Paul Blocq, writes in the *Traité de Médecine*, published by Charcot, Bouchard, and Brissaud, on the "Theorie Infectieuse" in chorea, as follows:-

"La chorée survient fréquemment à la suite des maladies infectieuses; elle s'accompagne souvent d'arthropathies, parfois de fièvre, d'endocardites, de suppurations; elle est sujette aux récidives, et donne lieu en certains cas à des troubles mentaux; ce sont là autant d'anal-

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-ogies qu'elle a avec les maladies infectieuses, analogies qui ont été invoquées à l'appui de la théorie infectieuses.*****Pour certains auteurs, la chorée est une infection spécifique reconnaissant pour cause un microbe special. Pour d'autres, elle tire son origin d'agents infectieux divers, qui la réalisent en vertu d'une prédisposition individuelle. Il va de soi qu'en l'un comme cas se pose la question subsidiaire de savoir si les manifestations choréiques sont liées à la présence des microbes eux-mêmes (infection) ou de leurs produits de sécrétion (intoxication)." (Traité de Médecine, p. 1216 & 7.)

M. Triboulet, who has done an immense amount of work on this subject, and wrote a noted thesis on the subject in 1893, writes in the "Traite de Medecine", published under the direction of MM. Brouardell and Gilbert, Paris 1902, page 389, as follows-

"La théorie microbienne spécifique n'est pas prouvée bactériologiquement; elle contredit le fait de la

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variabilitié étiologique,variabilitié confirmée,du moins en apparence,par la bactériologie.*****

La plupart des auteurs n'ont pas recontré les microbes in situ,ni sur les centres encéphaliques,ni sur la moelle,ni sur les nerfs. Les troubles nerveux choréïques apparaissent alors comme résultant d'une intoxication,supposition que ne combat aucune donnée positive,et que les recherches physiologiques actuelles sue les intoxications nerveuses produites par les poisons microbiens viennent pleinement corroborer."

In 1888 Naunyn demonstrated in a case of chorea,the presence of a cladothrix in the meningeal haemorrhages. Stein Kopff in 1890,in a case of chorea,demonstrated the presence of staphylococci, and streptococci, in the nervous system, and in the endocarditis.

Mircoli in 1892,established in certain cases of chorea, the presence of staphylococci and streptococci in the nervous system.

H.Meyer of Bale in 1894,found both staphylococci and streptococci in the blood and in the brain of a choreic

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patient.

Dr. Dana, (New York Med. Journal Aug. 1893) reports a case of microbic origin, a man of 26 years of age who had suffered from the disease from the age of 15. The attacks were similar to those of the ordinary Sydenham Chorea. At first there were intermissions, but the disease finally became chronic, and the patient died from exhaustion. There was no heart disease or rheumatism. The post mortem showed a conspicuous chronic lepto-meningitis involving the vertex of the brain. Microscopically, this was found to be mainly a proliferation process, without exudation or much cell infiltration. In the superior layer of the cortex there was cell infiltration with degenerative changes. In this region and in the deep layer of the pia a diplococcus was found. In addition there were found hyaline bodies in the outer layers of the cortex, and less numerically in the basal ganglia. There were evidences of meningeal irritation, vascular disease (arteritis), and nerve root irritation in the medulla and upper part of the cord.

Berkley reports a case where cultures were made from

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the blood of the left ventricle, from the vegetations on the valves of the heart, and from the parotid gland of a choreic patient, showing an abundant growth of the staphylococcus aureus.

Triboulet, (Paris Thesis 1893) has made a number of observations on the infective nature of the disease at the L'Hôpital Des Enfants, Paris, which he describes in this thesis. He made a large number of cultures from two fatal cases, finding staphylococci. In another he found the same organism in the blood during the febrile attack. He also investigated the condition of the spinal cord in a number of dogs suffering from canine chorea, and found marked lesions of the cells of the anterior horns. Wood, ("Nervous Diseases and their Diagnosis"), has described similar changes. Both however are agreed that canine chorea is quite a different disease to that seen in man.

Pianese of the Pathological Institute of the University of Naples has made an exhaustive research on the subject. He describes a bacillus which he has isolated from the

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nervous system of a choreic patient, and from which he was able to make cultivations. Inoculations into animals produced death with muscular twitchings, and convulsions, and from these animals he obtained pure cultures of the same bacilli from the central nervous system. (La Natura Infettiva della Corea del Sydenham, Naples, 1893). Duchateau in his "Essai De Pathogénie De La Chorée De Sydenham, Paris 1893", ^(page 43.) describes these experiments and their results as follows:-

" Les recherches ont été faites à l'aide de bacilles trouvés dans la cervelle cervicale d'un choréique et cultivés sur gélatine à la temperature de 20° à 38° C. Les cultures ont été inoculées sans succès sous la peau, dans la péritoine et dans les veines chez des lapins, cobayes et chiens. Les résultats ont été plus heureux par l'inoculation dans la dure-mère, la moelle, le nerf sciatique du chien et du lapin et sous la muqueuse nasale du cobaye. Les phénomènes ainsi provoqués consistaient d'abord en tremblements musculaires parfois généralisés et parfois localisés à quelques

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groupes de muscles, en particulier à ceux de l'épaule et de la région fessière; et dans une excitabilité extrême de l'animal, puis après quelques jours en contractures avec impotence partielle des membres. Alors l'animal succombait vers le 4^e Jour. Des chiens et des lapins qui survécurent, présentèrent des phénomènes pendant 20 à 30 jours, chez ceux qui moururent on trouva dans les centres nerveux, des bacilles que l'on put cultiver et qui étaient identiques à ceux des cultures qui avaient servi aux inoculations." (page 43)

MM. Courmont and Rodet, (Bull. Soc. Biol. 28 Jan. 1892, and Traité de Médecine et de Therapeutique published by Brouardel & Gilbert, Paris 1902, "Chorée"), have made experiments on the toxines of the staphylococci pyogenes. When inoculated into dogs and rabbits they produced dyspnoea, an elevation of the blood pressure, and "excitabilité exagérée du système nerveux", muscular tremblings and some choreic movements.

Quite recently Beaton and Ainley Walker, have published a preliminary communication as to their researches on

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the microbial origin of acute rheumatism, and its allied conditions, (British Med. Journal Jan. 31st. 1903), they state as follows;—"We are now able to state, as the results of our own observations on the subject, that we agree entirely with the assertion that a micrococcus is constantly associated with acute rheumatic lesions, and is the causal agent in their production. And we believe the coccus which we have isolated from rheumatic cases to be identical with that obtained by Triboulet, Wassermann, Paine, Poynton, Meyer and several others:***** It may be added that the same micro-organism is obtainable from cases of chorea, and on injection into animals may produce a typical attack of acute rheumatism, just as do cultures from rheumatic cases. Up to the present time we have obtained the micro-organism from some 15 cases-viz. from 8 cases of acute rheumatism, 3 cases of chorea, and 4 cases of acute endocarditis in rheumatic subjects."

The general nature of the chorea cases examined and the particular source from which the coccus was obtained in each are shown, in the following table.--

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Case.	Source of Culture.	Remarks.
*****	*****	*****
9. Chorea, with heart failure.	Post Mortem heart's blood.	Inoculations made in rabbits with positive results.
10. Chorea.	Urine.	Ditto.
11. Chorea, heart failure.	P.M. heart's blood.	---

In this report they describe the micro-organism which they have called, "micrococcus rheumaticus".

With regard to the inoculations in the chorea cases they say as follows.—"We have been unable up to the present to see a true chorea result from our inoculations into the blood stream, although a number of quite young animals have been used. Our cultures, even from chorea in man, have up to the present given us only acute rheumatism in animals. On one occasion, however, we have observed violent spasmodic movements of the head upon the second day after inoculation, in an animal which had developed a mono-arthritis within 24 hours after its inoculation,

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and died on the third day with beady vegetations on the mitral valve;and in another animal,which suffered from polyarthrititis,and eventually recovered completely there was for several days paralysis of both hind limbs.*****Accordingly,we conclude that the bacterial specificity of acute rheumatism has been satisfactorily established.Its toxic specificity remains to be investigated;and is at present under examination".

On the one hand ,the course of the milder forms of the disease,and not infrequent association with emotional symptoms tends to the view that the disease is a neurosis;on the other hand the course of the severer forms,the arthrititis,the almost constant presence of endocarditis,speak for an acute infection.

The Auto-Intoxication Theory.

The subject of auto-intoxication has of late been considered, on the continent, more particularly, as a possible cause of some cases of chorea; owing to the fact that certain other obscure diseases have been proved as due to such a cause. Whether due to the products of micro-organisms introduced into the body, or due to a toxic action of the proteid products of metabolism produced in the body itself, it is a theory that has to be reckoned with in the study of the causation of this obscure disease. Although there are cases where the rheumatic theory of the disease would seem to explain the cause of it, others where the infective theory would explain all, there are others where, neither of these theories would do so.

Bouchard has demonstrated that many of the proteid products of metabolism possess a distinct toxic action. This is a distinct analogy between the action of animal tissues and the tissues of plants and bacteria. As to the latter, Pasteur has demonstrated the part played by them in disease and its production. He has found that, in many cases,

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bacteria relied almost entirely upon their poisonous products to help them in maintaining their existence in the higher animals. Bouchard has found that certain other of these proteid metabolic products have an antidotal effect on the poisonous products, especially when produced and acting in small quantities. These two sets of products differ markedly in their physiological activity. They may even ^{be} produced in the same cell, yet are characterised by perfectly distinct activities. Bouchard maintains that in the animal, and in the human body, even under normal healthy conditions, there is a constant production of poisonous proteid substances, which are constantly neutralised by antitoxic substances, equally the products of these cells; these products, moreover, he pointed out, are stored up in the blood to be called into use as occasion requires. (*Trans. of the Hunterian Soc.*, '95-'96. *Since Woodhead*, p. 89.)

Now these products of metabolism are eliminated by the various organs of excretion. If from any cause these excretory organs fail, there is an accumulation of poisonous products, too great to be neutralised, and

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therefore a toxic action is produced on the nervous system. This is well seen in the various nervous complications we get in chronic diseases of the kidneys, with deficient excretion of the urinary products; in the headaches resulting from constipation, etc. In addition we have these products removed by oxidisation with the oxygen in the blood. In excessive exertion, mental, or physical, we have an increased production of these products, and if the elimination of these products is interfered with, and an acculuration taking place, we have the nervous system particularly attacked. Now we know that at the age when chorea is most common, the growth of the child is most active, there is great activity in the tissues, along with this we have an increase of these poisonous proteid products of metabolism. In a great number of choreic patients we find they are in a debilitated condition from various causes where there is likely to be defective elimination, and oxidation of waste products; they therefore accumulate in the body. It is not unreasonable to suppose, as no distinctive cause of the disease has been described, that

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a toxic condition may be developed which acts specially on that portion of the nervous system concerned in chorea, and that in its action, it is an irritant, gradually wearing out the patient by exhaustion, or by intercurrent complications in those cases that prove fatal. The hypothesis is not an unlikely one, in effect how much do we not see of affections of this nature, eclampsia, uraemia, and other diseases. MM. Boinet, and Silbert, (Rev. Med. Jan. 1892), have studied the principal agents contained in the products of the evaporation of the urine in these diseases, and have found some with energetic convulsing properties, so much so that small doses injected into rabbits, caused death. By dissolving these products, they were able to reproduce by inoculation all the symptoms of the disease.

Dr. R. Lépine of Lyons has made experiments on auto-intoxication by the kidneys. He introduced into the ureter of a dog a cannula communicating with a reservoir of sterilised water, to which 0.7% of chloride of sodium was added. The reservoir was sufficiently elev-

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-ated to prevent the exit of the urine, and to ensure the entrance into the kidneys of a small quantity of the saline solution. The following phenomena occurred; the animal foamed at the mouth, the central and peripheral temperature rose gradually and almost simultaneously, the respiration became slower at first, then became quicker and very noisy; there were occasional jerky movements of the paws; the central temperature continued to rise, and in a few hours the animal died, with a temperature that varied from 104° to 107.6° F.

A considerable quantity of sterilised salt water may be introduced into the veins of a dog without causing any notable disturbance. The symptoms described cannot therefore, be attributed to the mere introduction of the water. Being introduced through the urinary passage, however, the water washes the kidney before entering into the circulation, and carries with it the interstitial juices of the kidney; thus exercising a thermogenic and dyspnoeogenic action on the organ. From the result of this and other experiments Dr. Lépine

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concludes that the healthy kidney contains substances having thermogenic and dyspnoeogenic properties.

In addition to the various poisonous animal alkaloids which are produced in the body in health, there are others which have been isolated during disease, and when injected into animals have caused either a similar disease or death from some other cause. Pouchet has extracted an alkaloid from the excretions of cholera patients, which when injected into animals, caused slowing of the heart and then death, followed quickly by rigor mortis. Pouchet, Nicati, and Rietsch, have obtained an alkaloid from Koch's tubercle bacilli, which from experiments seem to be of a similar nature. Brieger, from cultivations of the tetanus bacilli, has extracted four ptomaines, all of which when injected into mice caused death by tetanus. In the case of tetanus, and diphtheria, we know now, that it is the products of micro-organisms that the disease in the human subject is due to.

We see in these diseases mentioned and also in such diseases as septicaemia, uraemia, and similar conditions

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these poisonous products seem to have a special affinity to attack the nervous system.

Among certain children who are not suffering from any degenerative diathesis, where the disease shows itself at the period of most active growth, it is a reasonable hypothesis. In cases where there is already a predisposition, and the patient debilitated from anaemia, rickets, rheumatism and other similar diseases, the intoxication will it is reasonable to suppose act more easily on the already exhausted nervous system.

The chorea of pregnant women we place as likely to be due to a process of intoxication, the excretory functions, and the process of assimilation being often at this time, at fault. Although at this time the chorea is most usually ascribed to some fright, that the patient has received ; it is possible that the emotion produced by the sudden shock was a pathological consummation of stimuli that was required to start the disease in the nervous system already exhausted by the ever increasing toxic poisons. The rapid recovery after delivery when the whole system finds relief in the various discharges

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that then take place is certainly in favour of such a theory.

It seems then not unlikely that some cases of chorea may be due to a process of intoxication due to abnormal changes, independent or dependent on a bacterial agency, which may result in the formation of poisons having a special affinity for the nervous system.

The complex chemical history of the body in disease is yet in its infancy, and it seems that in the future we shall find the cause of many now obscure diseases in the chemistry of the body itself.

In chorea where the pathology has only taught us negative facts it is necessary to bear in mind the possible theory of auto-intoxication as a cause of the condition in some cases.

11. CONCLUSIONS.

In coming to a determination as to the exact cause or causes of this affection, one has to consider the following points.--

1st. What the clinical study of the disease has taught us.

2nd. What the study of the Morbid Anatomy, and Pathology, has taught us.

First. The fact that it is a disease pre-eminently of childhood, and the time of puberty, and tending as a general rule to recovery, and knowing how common it is to find at this time the child attacked by various infectious diseases, leads one to look at the infective theory for a cause.

In the light of present discoveries in connection with Acute Rheumatism, with which it seems in a certain percentage of cases to be associated; one is again brought face to face with the infective theory.

Just as in rheumatic cases we have many types of rheumatism, so we have many types of chorea. Certainly in all cases it seems difficult to imagine that the disease

(Conclusion.)

is of microbial origin, there are however a certain percentage of cases where it seems to be the only explanation. So it is in rheumatism; although the acute rheumatic fever, has all the symptoms and complications of an acute infective process, the milder cases with slight fever, with little constitutional disturbance, are difficult to ascribe to an acute infective process. The severe fatal cases of chorea, with fever, endocarditis, delirium, mania, etc. one is able to consider them as part of an acute infective process. The mild cases without constitutional disturbance, without fever, without complications it is difficult so to do. The probability is that as our methods of research become developed we shall find that in these two diseases, we have subdivisions due to entirely different causes. Just as we have found in Phthisis, that some cases are due primarily ~~due~~ to the tubercle bacilli, others due in the first instance to irritation, as in the case of the stone mason, the coal miner, etc. or due to an infective pneumonia. The great cause, predisposition, being there in

(Conclusion.)

all cases, no matter what the exciting cause is. So with chorea, and rheumatism, we shall probably find that the severe cases, with their serious complications are due to specific microbes or their toxins, distinct but allied to each other. While others may be due to a process of intoxication, that goes on in the body, from a disorganisation of nutrition, and elimination, whereby certain as yet unknown products are formed which act as an irritant to the nervous system in the case of chorea. Such is in all probability the condition amongst those cases we get in pregnancy with no neurotic or rheumatic history. Also in many children with healthy surroundings, and no degenerative history. Many who are opposed to the infective theory of the disease, argue that the psychical element in the disease is too strong to admit of it having an infective origin. But when you consider how common this is in almost all children, and especially in neurotic ones, it is not surprising to find a history of it in this disease,

(Conclusions.)

which affects the nervous system in such a manner.

It is simply the exciting cause to an already exhausted nervous system;the last pathological straw.

It is true that some cases seem to be allied to the condition known as hysteria,and have been classed on that account as neuroses.That condition itself is,however little understood,and its pathology still remains obscure.

Secondly. In studying the Morbid Anatomy,and Pathology, of Chorea,we get little help except of a negative character.In the various conditions and already referred to,we find no characteristic lesions present in the nervous system,to which we can attribute the disease.This when we consider the striking,and unmistakable symptoms of the disease,is remarkable.The cases where it has been possible to make an autopsy, have all been acute cases,and the conditions found have been such,that they can all be looked upon rather as the result of the disease and not the cause.

(Conclusion.)

We have another disease of the nervous system, which is undoubtedly due to a specific micro-organism, and which is directly comparable to many cases of chorea, viz. Epidemic Cerebro-Spinal Meningitis. Here in a variable percentage of cases we have acute arthritis, just as we see in some cases of Chorea. Flexner and Barker, (American Journal of Medical Sciences 1894), report an epidemic of Cerebro-Spinal Meningitis, with 20% of the cases complicated with acute joint affections, resembling acute rheumatism. There is scarcely an argument used by the strong advocates of the rheumatic nature of chorea, which could not be forcibly applied in favour of the rheumatic nature of epidemic cerebro-spinal meningitis.

The fact that complete recovery is the rule, and not the exception, without leaving any after effects in those cases that have had no complications, seems to favour the infective theory of the disease. Just as we have no after effects in the majority of the infective diseases of childhood without complications. Looking at the diseases

(Conclusions.)

of the nervous system classed as Neuroses we rarely find so complete a recovery. Just as we have various forms of Epilepsy, with different causes, so we have Choreas with different causes, some due to an intoxication, caused by a defective metabolism and elimination at a period when growth is at its height, some due to a condition of the nervous system allied to hysteria, others due to a specific micro-organism, or its toxins, as we get in tetanus, and diphtheria.

"Étiologiquement il y a plusieurs chorées comme il y a plusieurs épilepsies".

***** " *****

12. NOTES ON CASES ATTENDED, PERSONALLY.

Case 1. Ellen B. age 18. 1st. attack. Family history good except mother suffered from "rheumatic pains". General surroundings good. Patient healthy and well nourished, never had rheumatism. Measles when 3 years old. 2nd. week of attack which was confined to face and left arm, Erythema Nodosum appeared on the anterior surface of the left leg, which disappeared in a few days. No fever no complications. Termination. Recovery. No complications.

Case 2. Mrs. W. age 42. Had chorea for 10 years. I was called in on account of her having broken left collar bone, the result of a fall. Family History. - Father dead, cause unknown, suffered for many years from Megrin. Mother died some years ago from "heart disease". Married life very unhappy; had 5 children, no rheumatism. Present attack came on 10 years ago just before her last child was born. Attack attributed to being pushed downstairs by her husband. Present Condition, inco-ordinate movements general, but worst in the arms. Speech affected. Mind not affected. No heart complications. Apart from the chorea, health fairly good. Been under various medical men in London during the last ten years, but no treatment seemed to affect the chorea. Fracture of the clavicle united, after considerable difficulty owing to the choreic movements.

Case 3. Alice H. age 18. Attack commenced a week before I saw her. Family History. Father and Mother healthy. Four brothers all healthy. One sister suffered from tubercular glands in the neck. (Which I subsequently operated on), and

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her eldest sister was somewhat dull mentally.

Patient ^{was} ~~is~~ at the commencement of the eight month of her first pregnancy, she has always lived in the country amidst healthy surroundings, looks healthy and well nourished. She had a slight attack of chorea when 5 years old, which lasted for three weeks. Never had any rheumatism. Heart normal. When first seen the choreic movements were confined to the arms and face. Towards the beginning of the 9th. month the choreic movements became general, speech was affected, temperature, 102. pulse 120, and her general condition very serious. Chloroform had to be administered frequently, and on two occasions she became distinctly maniacal. Induction of labour was recommended on two occasions, but the parents objected. Pregnancy went on to full time notwithstanding the violence of the choreic movements. Chloroform was administered during the whole of labour at intervals. The choreic movements gradually subsided after the confinement; but it was six weeks before they had entirely disappeared. This attack was attributed to fright by the parents. On investigation of this point, I found the fright she had had was a week previous to the onset of the attack. This patient has gone through another pregnancy since without any signs of chorea, Patient is now in good health, heart normal.

(Since writing above I have heard, that this patient is approaching her 3rd confinement, & is suffering from violent Chorea). D.K.

Case 4. Fred W. age 7. Admitted into H.--Workhouse, suffering from Hemi-chorea of Right side. No family history obtainable. Child badly nourished, and anaemic. No indications of rheumatism, heart normal. Pulse 100, temp. 99. Treated by rest in bed, and generous diet only. Termination. Complete recovery in three weeks.

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Case 5. Jane J. age 7 years. Family History. Father had rheumatic fever. Mother alcoholic. Brothers and sisters healthy. Had "fits" when a baby, and had scarlet fever six months previous to attack of chorea. Inco-ordinate movements of head arms and slightly in legs. Heart normal. Pulse 90, temp. sub-normal. Termination; complete recovery.

Case 6. Tom. J. age 4, brother of above patient. This boy as he was suffering from a bad cold was confined in the same room with his sister who had chorea. He also had had scarlet fever six months previously; but was an exceedingly healthy child, with a very passionate temper. Three days after being confined with his sister he developed choreic movements in the face and right arm. They appeared to be quite uncontrollable. He was isolated in another room, and in less than a week the movements disappeared without any other treatment. (I looked upon this case as hysteroidal in nature, notwithstanding the age of the patient.)

Case 7. Henry G. age 20. Family History, father strong healthy farmer but inclined to be alcoholic. Mother dead some three years of "heart disease". One sister healthy. Had two previous attacks of rheumatic fever, (during which attacks I attended him). Present attack of chorea commenced during the second week of his third attack of rheumatic fever, and during an attack of endocarditis. The choreic movements were confined to the face muscles and the speech was also affected. There were occasional twitchings of the right arm; but they were always under control. This patient was very seriously ill for some six weeks, but subsequently recovered. The chorea however left him with a mitral systolic

(Notes on Cases.)

murmur, and the speech is still affected to a certain extent five years after the attack.

Case 8. Ann G. age 12. Had been ill for two weeks with pains in the joints and slight movements of the arms before I saw her. No family history obtainable. Patient seemed weak and ill nourished, and decidedly anaemic. When seen choreic movements were developed in both arms and in the muscles of the face. Temp. 103, pulse 120. The patient subsequently had pericarditis. She was treated with Sodium Salicylate and blisters over the heart. Termination, Recovery, with a heart murmur over apex.

Case 9. May W. age 14. 2nd. attack. Family History, Father healthy but subject to Quinsy, mother dyspeptic and distinctly neurotic. Brothers and sisters healthy. Has had measles, and scarlet fever, and rheumatic fever three years ago. I was called to attend this patient for a second attack of rheumatic fever,; she had a loud systolic murmur at the apex, and joint pains, in the knees, ankles, and shoulders. Pulse small, regular, 120, temp. 103. She suffered from very severe pains over the heart, like angina, frequently. It was after one of these attacks of angina, that choreic movements began in the face muscles and in the left arm. At the same time the joint pains left her, nor did they again return to any extent, although the angina pains continued at intervals for about two weeks; but were always relieved by blistering. The choreic movements were never severe and gradually ceased as the patient became convalescent. She was treated with anti-rheumatics, rest, blistering and a little opium when the heart pain was severe. She very slowly recovered, and was left with a loud mitral

(Notes on Cases.)

systolic murmur. This child was an extremely bright and intelligent one, and had always lived at the sea-side, in a good home.

Case 10. Wilfrid B. age 10. Strong healthy boy. Family history. Father healthy, mother had rheumatic fever when a girl. Patient had scarlet fever 4 years before attack, and was also operated on for adenoids directly after. Choreic movements general, and severe. No rheumatism or other complications. Treated by rest in bed and large doses of arsenic. Complete recovery in about a month. Heart not affected. This child had a very good home, and had not received any fright or other exciting cause as far as one could tell.

Case 11. Violet D. age 15. Was called to see this patient on account of continued violent headache in the frontal region. Family History, -father had suffered from headache all his life, mother was distinctly neurotic, but otherwise healthy. Two brothers healthy. Patient had suffered from headache when at school, "off and on", for some time, This attack however was the worst she had had. On examination, all systems seemed normal with the exception of the reproductive where there had been considerable pain at the menstrual periods. The headache seemed ⁴ little affected by treatment; the eyes were then examined with the ophthalmoscope. What was diagnosed as optic neuritis was discovered. The patient was confined in a dark room, the temples were blistered and bromides administered. She gradually recovered, but developed choreic movements in the face muscles, and in the arms. She was still confined to bed and chloral hydrate added to her medicine. She made a slow recovery,

(Notes on Cases)

and had no return of the headaches.

Case 12. George I. age 5. Was called to see this patient on account of a severe fall which happened some 3 hours before I saw him. Family History and surroundings good. No history of any previous illness. I had attended the mother when the child was born, and had never known anything the matter with it. The patient was said to have been unconscious for some time after the fall. When I saw him there were choreic movements of the arms, and occasionally of the head. Cold in the form of ice was applied to the head. The child was kept in bed and purged with calomel. The general condition improved but the choreic movements continued, and the child was then put on large doses of arsenic. Recovery quickly took place.

Note. This certainly was a very interesting case, whether the fall had anything to do with the choreic condition, or was it only a coincident? If there had been a blood clot pressing on the motor centres one would have expected only one side to be affected. It is hardly conceivable that both sides of the brain would be so affected. Again it is unlikely that there could have been a similar condition lower down in the motor tract without causing other symptoms.

I found after that the child had been ailing slightly for some little time and had been very, "cross and irritable". I therefore came to the conclusion that the attack of chorea was independent of the fall which was only a co-incident.

(Notes on Cases)

Case 13.

David G. age 16, Healthy young lad, never had rheumatism or chorea previously. No cause for present attack given. Family history and surroundings good.

The choreic movements involved the limbs and the face muscles. The tongue was protruded with a jerk. The mental power was certainly affected. Heart normal, Urine, acid, sp. gr. 1030, no albumen. Mental condition gradually got worse. He was treated with arsenic; but did not improve. Two weeks after the onset of the chorea, the temperature went up to 102, and he complained of pain in the praecordial region. There was increased action of the heart and a pericardial murmur was made out. The choreic movements became more severe. The next day there was swelling of the knee and ankle joints which were also very painful. There was delirium at night, and the pulse went up to 130. The patient was put on Salicylate of soda and a blister applied over the heart. This treatment greatly relieved the pain over the heart which at times had been very severe, the temperature also came down to 100. The rheumatism gradually left him, the friction sounds over the heart also disappeared, and the choreic movements subsided. The mental power also became much improved.

This case certainly seemed of infective origin. The pericarditis and rheumatism did not appear until two weeks after the onset. On the appearance of the pericarditis the choreic movements were increased, and the general condition became very much worse. On the fever declining the chorea at once became less severe, and

(Notes on Cases.)

the mental condition began to improve. This patient was seen by me some three years after, and seemed in perfect health, the heart was normal, and the mental condition quite recovered.

Case 14. Phyllis G. age 7. Slight choreic movements of face and right arm. No rheumatism, other systems all normal. Family history, father gouty, but otherwise had very good health. Mother healthy, but certainly inclined to be neurotic. No cause for present attack. Child had had measles some two years previously. General life and surroundings everything that could be desired. Treated by rest and arsenic. Perfect recovery, no heart complications.

N.B. The above cases were all attended in private practice and led me to study the cause of this interesting disease. The notes on these cases were not taken with any idea of writing a thesis on the subject.

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